J Exp Neurol. 2024;5(3):162-166.

Journal of Experimental Neurology Case Report

Acute Obstructive Hydrocephalus and Acute Cerebellar Ischemic Stroke as a Complication of Takayasu Arteritis in a Young Male Patient: A Rare Case Report

Muhammad Hashim[1,#], Asif Khan[1], Muhammad Khadim[1], Sabir Shah[1], Awais Ahmad[1], Farhan Shahzad[2,#,*]

[1]Department of Medicine, Khyber Teaching Hospital, Peshawar, Pakistan

[2]Saidu Group of Teaching Hospital, Swat, Pakistan

[#]Equally contributed

[*]Correspondence should be addressed to Farhan Shahzad, Farhanshahzad09@gmail.com

Received date: July 09, 2024, **Accepted date:** August 29, 2024

Citation: Hashim M, Khan A, Khadim M, Shah S, Ahmad A, Shahzad F. Acute Obstructive Hydrocephalus and Acute Cerebellar Ischemic Stroke as a Complication of Takayasu Arteritis in a Young Male Patient: A Rare Case Report. J Exp Neurol. 2024;5(3):162-166.

Abstract

Introduction: Takayasu arteritis is a rare type of vasculitis associated with various neurological manifestations. Multiple components, including thrombosis, embolism, stenosis, and vascular inflammation, are involved in the underlying pathophysiology.

Case Presentation: A 21-year-old male patient presented with vomiting, difficulty speaking, disorientation, and seizures. On examination, pinpoint pupils and blood pressure discrepancies among four limbs were noted. Imaging findings supported the diagnosis of acute obstructive hydrocephalus, acute cerebellar infarction, and Takayasu arteritis. The patient was treated with IV antibiotics and steroids and underwent a ventriculoperitoneal shunt.

Case discussion: Takayasu arteritis with cerebellar ischemia and obstructive hydrocephalus is quite a rare case. It presents with headache, dizziness, disorientation, and rarely with stroke. The diagnostic process includes computed tomography angiography (CTA) and imaging of the brain. It is mostly treated with pharmacological therapy; however, in some cases, surgery is required.

Conclusion: This case highlights the rare but serious neurological adverse effects of Takayasu arteritis in young male patients, as well as the importance of interdisciplinary care. To prevent neurological degeneration, Takayasu arteritis patients must be identified and treated as soon as possible.

Keywords: Cerebellum, ischemia, hydrocephalus, peritoneal shunting, Takayasu arteritis, CT angiography, young male patient

Introduction

Compared to other types of vasculitis, Takayasu arteritis, a rare disease, is more likely to cause neurological symptoms such as headaches, dizziness, stroke, and transient ischemic attacks [1]. The underlying pathophysiology involves multiple components, including thrombosis, embolism, stenosis, and vascular inflammation [1]. The best method for diagnosing Takayasu arteritis is computed tomography angiography (CTA) [2]. Treatment options include pharmacological therapy, endovascular procedures, and, if required, open bypass surgery [2]. This case report discusses a rare instance of Takayasu arteritis that could have caused acute cerebellar ischemic stroke and acute obstructive hydrocephalus.

Case Presentation

The primary complaints of a 21-year-old male patient, who was referred from a primary care hospital with acute disorientation, vomiting, slurred speech,

lethargy, and focal seizures affecting the upper limbs. The patient appeared drowsy upon examination, and his blood pressure in the upper limbs measured 130/95 mm Hg in the right upper limb and non-measurable (unrecordable) in the left upper limb, while it was 155/110 mm Hg in the lower limbs. Pulse was 110 beats per minute in the right upper limb but non-palpable in the left upper limb. Oxygen saturation was 93% on room air. Cardiovascular examination revealed normal first and second heart sounds with the presence of a carotid bruit over the right carotid artery. Abdominal examination was unremarkable. Neurological examination revealed bilateral pinpoint pupils unresponsive to light, with a Glasgow Coma Scale (GCS) score of 10/15 (E3V3M4), right plantar withdrawal, and mute left plantar reflexes.

The routine blood tests revealed a normal coagulation profile, and liver and kidney functions; however, urine analysis showed 8-10 pus cells and urine culture demonstrated heavy growth of *Acinetobacter* species sensitive only to tigecycline. Cerebrospinal fluid (CSF) analysis was unremarkable, and the remaining investigations can be found in **Table 1**.

An echocardiogram revealed no abnormalities, while a magnetic resonance imaging (MRI) showed findings consistent with acute ischemic infarction in the right cerebellum, pons, middle cerebellar peduncle, and right posterior temporal region (**Figures 1A** and **1B**) along with acute obstructive hydrocephalus (**Figures 2C** and **2D**).

Following consultation with the Department of Neurosurgery, a ventriculoperitoneal shunting procedure was performed under general anesthesia. Upon emergence from general anesthesia, the patient experienced difficulty maintaining adequate oxygen saturation, necessitating transfer to the intensive care unit (ICU) for ventilatory support. Subsequently, a tracheostomy was performed to facilitate prolonged

Table 1. Investigations.

S. No	Test(s) Name	Result(s)	Normal Range(s)/Unit(s)	Comment(s)
1	Total Leukocyte Count (TLC)	16×10^3	$4.5\text{-}11\times10^3$/uL	Elevated
2	Hemoglobin	14	13-17.5 g/dL	Normal
3	Platelet	275×10^3	$150\text{-}450\times10^3$/uL	Normal
4	Serum albumin	3	3.5 and 5.0 g/dL	Low
5	Creatine kinase (CK)	547	22-194 U/L	Elevated
6	Creatine kinase-MB (CK-MB)	57	5 to 25 IU/L	Elevated
7	Lactate dehydrogenase (LDH)	601	120-240 U/L	Elevated
8	C-reactive protein (CRP)	101	less than 5 mg/L	Elevated

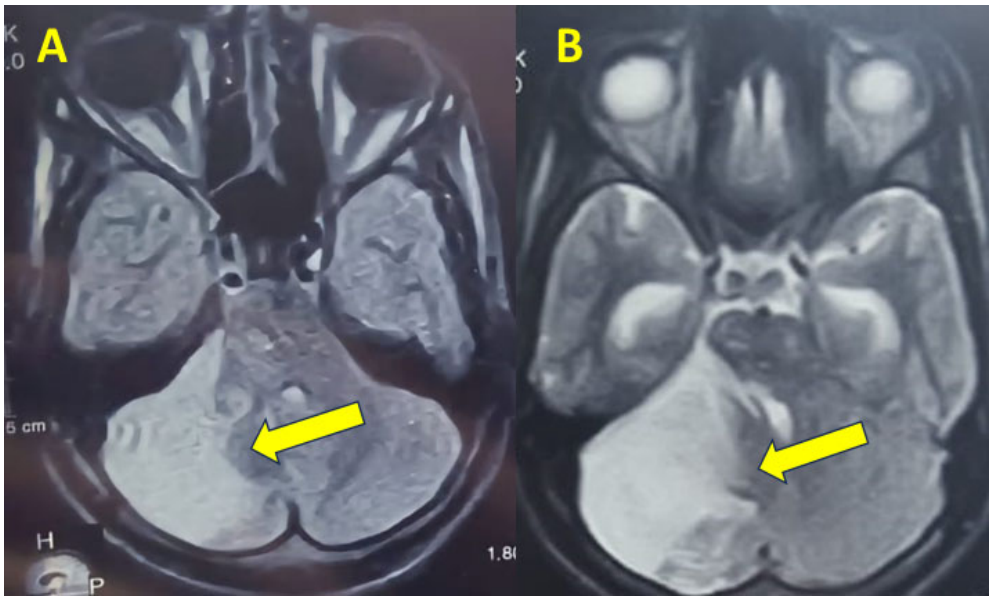

Figure 1. T2WI; Ischemic stroke (yellow arrows in images **A** and **B**).

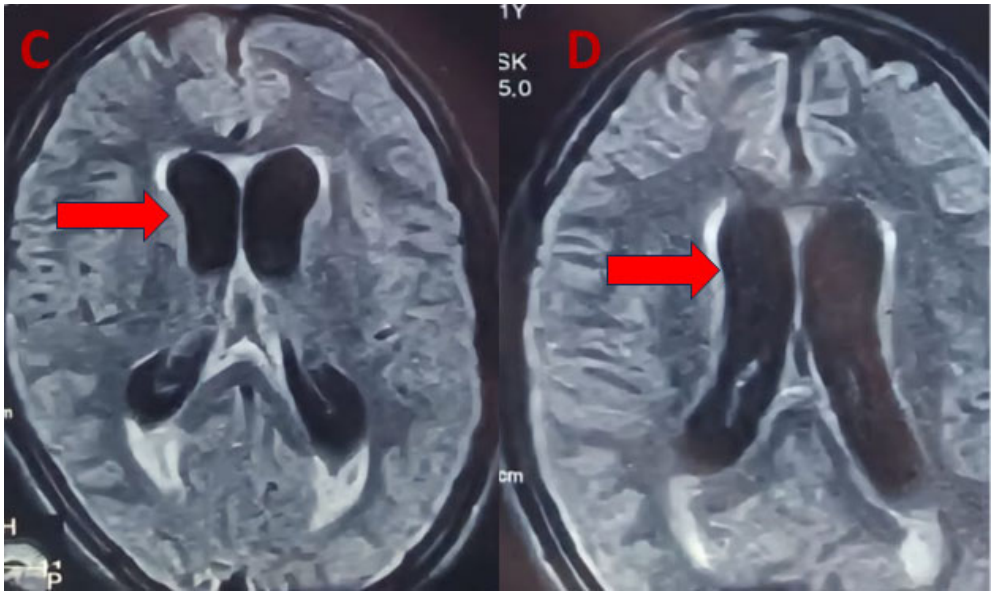

Figure 2. Flair with squashed CSF signals; Hydrocephalus (red arrows in images **C** and **D**).

ventilatory assistance. He was extubated after three days. CTA revealed vasculitis of the arch of the aorta and its major branches suggestive of Takayasu arteritis type IIa (**Figure 3**).

Takayasu arteritis was diagnosed using the American College of Rheumatology (ACR) criteria in which 5 out 6 criteria were present in this patient (**Table 2**).

He was put on broad-spectrum intravenous (IV) antibiotics, initially meropenem, which was changed to tigecycline based on urine culture and sensitivity. IV linezolid, IV omeprazole, IV multivitamins, IV Dexamethasone, IV Valproic acid, IV levetiracetam, Beclomethasone, and ipratropium nebulization were further included in the treatment.

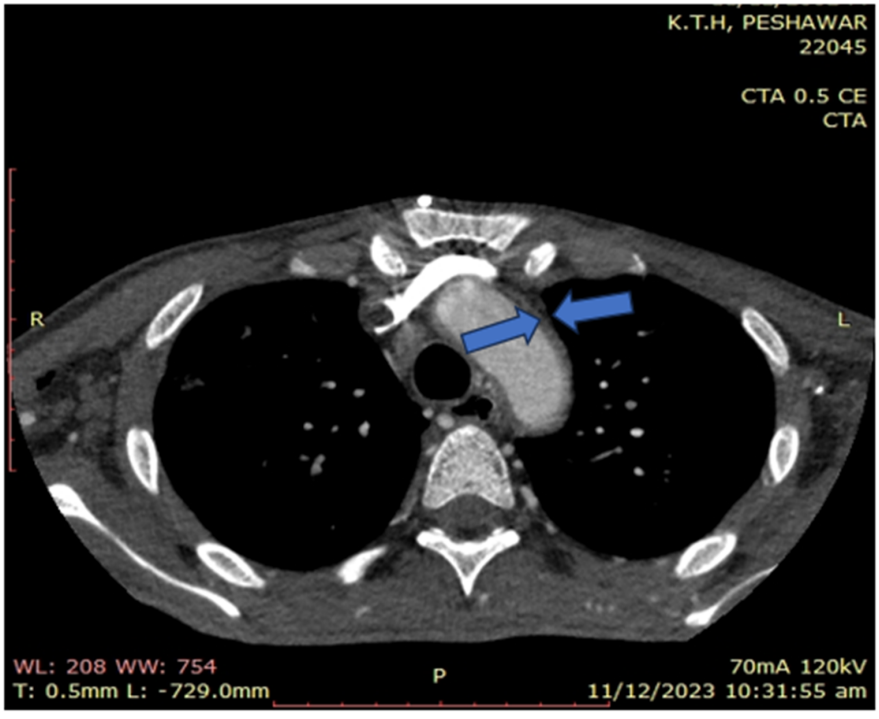

Figure 3. CTA showing thickening of wall of arch of aorta (blue) suggestive of Takayasu arteritis.

Table 2. ACR diagnostic criteria for Takayasu Arteritis.	
ACR criteria	**Criteria present or not in patient**
Age at Onset <40 years	Yes
Systolic **blood pressure d**ifference >20 mmHg between arms	Yes
Claudication of **e**xtremities	No
Vascular **b**ruit	Yes
Decreased or absent **b**rachial **p**ulse	Yes
Arteriographic **a**bnormality	Yes
TOTAL: (3/6 ARE REQUIRED FOR DIAGNOSIS)	5/6

The patient recovered from hydrocephalus; however, he experienced persistent neurological deficits, such as motor weakness, coordination problems, and speech difficulties. He was discharged from the hospital and thereby referred to a Rehabilitation Centre for physical and speech therapy.

Case Discussion

Takayasu arteritis is a rare type of vasculitis, that can result in stenosis, occlusion, and aneurysm of large arteries predominantly in females and is more common in Asians [1] but in our case, it affected the male patient. More than half of patients with Takayasu arteritis can show neurological manifestations like dizziness (78%), headaches (25%), transient ischemic attack (3-22%), and stroke (10-20%); however, our patient did not demonstrate dizziness or headache but presented with the rare manifestation of stroke.

Patho-physiologically, it is a condition characterized by inflammatory granulomatous vasculitis affecting medium and large arteries. This inflammation can result in the thickening, fibrosis, and ultimately occlusion of multiple blood vessels and consequent ischemic manifestations. Furthermore, the ischemic stroke may be caused by aneurysm and sinus venous thrombosis, meningeal and brain parenchymal involvement resulting from granulomatosis and perivasculitis, and encephalopathy due to cytokine damage [2].

The diagnosis of Takayasu arteritis is frequently delayed due to its nonspecific clinical presentation. Takayasu arteritis is diagnosed based on Modified Ishikawa's criteria and the ACR criteria (**Table 2**) [3,4]. The treatment modalities for Takayasu arteritis include pharmacological therapy (the predominant modality used in our case), endovascular intervention, and open bypass surgery [4]. Commonly utilized agents encompass corticosteroids and traditional immunosuppressive medications, but for resistant or intolerant patients, biologic drugs such as tumor necrosis factor (TNF) inhibitors may be used. In cases of arterial stenosis, balloon angioplasty, stent graft replacement, or surgical bypass may offer benefits [5].

Strokes are less frequent in younger adults compared to older adults, with approximately 10-15% of all strokes occurring in individuals aged 18 to 50 years. Unexpectedly, this occurred in our study [6]. Vasculitis represents a rare etiology, accounting for 3-5% of strokes in individuals under the age of 50 [7], again highlighting the rarity of our study. Among individuals with TA, 59% experienced neurological impairment, 35% reported recurrent strokes, and 24% developed epilepsy [8].

Hydrocephalus in this case could be secondary to posterior fossa oedema caused by hypertensive posterior reversible encephalopathy syndrome [9,10]. It can emerge as a rare yet life-threatening complication of systemic autoinflammatory diseases, including primary vasculitis. To date, hydrocephalus has been reported only in a single case of TA, making our study a significant contribution to the medical literature. It is managed through anti-inflammatory or anti-infectious treatments, with surgical interventions such as ventriculoperitoneal shunts proving effective in the majority of cases [9,10].

In our study, we highlight a unique case of Takayasu arteritis in a young male patient who developed an acute cerebellar ischemic stroke and hydrocephalus as complications. Early diagnosis and management of Takayasu arteritis in young patients can prevent such complications from occurring.

Conclusion

In conclusion, this case report highlights the rare but significant complications of Takayasu arteritis in a young male patient, who presented with acute obstructive hydrocephalus and acute cerebellar ischemic stroke. The simultaneous occurrence of these complications underscores the complexity and diverse manifestations of this vascular disorder. This case serves as a reminder for healthcare providers to consider Takayasu arteritis as a differential diagnosis in young patients presenting with neurological symptoms. Prompt recognition and management are crucial to prevent further neurological deterioration and improve patient outcomes. Moreover, this case emphasizes the necessity of multidisciplinary collaboration involving rheumatologists, neurologists, and neurosurgeons in the comprehensive care of patients with Takayasu arteritis, particularly when encountering rare complications such as acute obstructive hydrocephalus and cerebellar ischemic stroke.

Declaration of Interest

No conflict of interests

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this Journal on request.

Ethical Approval

The study was approved by the Institutional Research and Ethical Review Board of Khyber Medical College Peshawar in accordance with the declaration of Helsinki (2013), IREB No: 367, Dated: 9th May 2024.

Funding Source

This research did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

Author Contribution

1. Writing the manuscript, Conceptualization: Muhammad Hashim (Main Author)
2. Review and editing: Asif Khan (Co-Author)
3. Writing the paper: Muhammad Khadim (Co-Author)
4. Data Curation: Sabir Shah (Co-Author)
5. Data Curation: Awais Ahmad (Co-Author)
6. Supervision, Proof Reading: Farhan Shahzad (Co-Author)

References

1. Zhang S, Yuan D, Tan G. Neurological Involvement in Primary Systemic Vasculitis. Front Neurol. 2019 Apr 26; 10:430.
2. Trinidad B, Surmachevska N, Lala V. Takayasu Arteritis. 2023 Aug 8. In: StatPearls [Internet]: StatPearls Publishing; 2023.
3. Sharma B, Jain S, Suri S, Numano F. Diagnostic criteria for Takayasu arteritis. International journal of cardiology. 1996; 54:S127-S33.
4. Bhandari S, Butt SRR, Ishfaq A, Attaallah MH, Ekhator C, Halappa Nagaraj R, et al. Pathophysiology, Diagnosis, and Management of Takayasu Arteritis: A Review of Current Advances. Cureus. 2023 Jul 29; 15(7):e42667.
5. Field K, Gharzai L, Bardeloza K, Houghton B. Takayasu arteritis presenting as embolic stroke. BMJ Case Rep. 2017 Aug 28; 2017:bcr2017220001.
6. Keser G, Direskeneli H, Aksu K. Management of Takayasu arteritis: a systematic review. Rheumatology (Oxford). 2014 May; 53(5):793-801.
7. Stack CA, Cole JW. The Clinical Approach to Stroke in Young Adults. In: Dehkharghani S, editor. Stroke [Internet]. Brisbane (AU): Exon Publications; 2021 Jun 18. pp. 53-78.
8. Mohammad AJ, Jacobsson LT, Mahr AD, Sturfelt G, Segelmark M. Prevalence of Wegener's granulomatosis, microscopic polyangiitis, polyarteritis nodosa and Churg-Strauss syndrome within a defined population in southern Sweden. Rheumatology (Oxford). 2007 Aug; 46(8):1329-37.
9. Tanna D, Mendiratta N, Negalur N. 9. An unusual case of Takayasu arteritis presenting as leptomeningitis with obstructive hydrocephalus. Rheumatology Advances in Practice. 2018; 2(suppl_1):rky033. 01.
10. Wei J, Yin H, Wang L, Cui L, Wang R. Systemic autoimmune diseases complicated with hydrocephalus: pathogenesis and management. Neurosurg Rev. 2019 Jun; 42(2):255-61.